British Heart Journal, 1978, **40**, 581–585

A case of surgically corrected Wolff-Parkinson-White syndrome
Clinical and histological data

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SUMMARY A case of type B Wolff-Parkinson-White syndrome, with intractable atrial fibrillation, underwent surgical division of a right-sided accessory atrioventricular bundle of Kent. Pre-excitation and complicating tachyarrhythmias were henceforth abolished for 6 weeks, when the patient died of infective endocarditis. Histological examination showed a divided Kent's accessory atrioventricular pathway and apparently functionless James and Mahaim fibres.

The surgical approach to management of intractable tachyarrhythmias in the Wolff-Parkinson-White (WPW) syndrome (Wolff *et al.*, 1930) relies upon correct assessment and permanent disruption of abnormal atrioventricular muscular connections (*British Medical Journal*, 1974).

Sophisticated electrophysiological investigations in the pre- and intraoperative stage (Durrer and Wellens, 1974; Gallagher *et al.*, 1975; Spurrell *et al.*, 1975; De Ambroggi *et al.*, 1976) are now enabling the surgeon to perform, in selected cases, a well-aimed division of anomalous atrioventricular bundles with encouraging results (Gallagher *et al.*, 1975).

However, much more information is still needed about the histological background of the WPW syndrome in general (Anderson *et al.*, 1975; Rossi, 1975), and in particular about its surgical features (Cole *et al.*, 1970; Iwa, 1975).

The present report describes a case of type B WPW syndrome, with atrial fibrillation, which was operated upon for division of a right-sided accessory atrioventricular bundle of Kent. Eventually, 6 weeks after complete surgical correction, histology made anatomoclinical evaluation possible.

Case report

A 47-year-old woman who had mitral and aortic valvular disease and type B WPW syndrome with paroxysmal tachyarrhythmias since 1965, entered hospital in 1974, when her rhythm disturbances increased, with signs of cardiac failure. The electrocardiogram (Fig. 1) showed shortened PQ interval (0·9 s) and delta wave widening the QRS complexes to 0·12 s, with predominant negative deflections (QS, rS) in right praecordial leads (Rosenbaum *et al.*, 1945). Recurrent attacks of atrial fibrillation were also recorded (Fig. 2), with delta waves in fast-rate QRS complexes (180 to 285/min) and with occasional bouts of ventricular fibrillation (Fig. 2). His bundle recording, in sinus rhythm (rate 64/min), showed AH interval of 160 ms and delta-H of 20 ms (Fig. 1). The impending threat of ventricular fibrillation discouraged atrial stimulation procedures.

An accessory atrioventricular bundle of Kent was suggested to be probably responsible for pre-excitation (Spurrell *et al.*, 1975) and for atrial fibrillation (Curry and Krikler, 1975), in which the impulses reached the ventricles downwards through the accessory atrioventricular pathway, at a fast rate (Fig. 2).

Oral ajmaline (300 mg/day), quinidine (1200 mg/day), and amiodarone (800 mg/day) failed to control the tachyarrhythmic attacks, which worsened, together with the patient's cardiovalvular conditions. Cardiac catheterisation confirmed aortic regurgitation and mitral stenosis with pulmonary hypertension, and it was decided to operate for double valve replacement and, possibly, for division of the Kent bundle.

Once the heart was exposed, anterior epicardial mapping was performed and early activation of the right ventricle identified, close to the lateral side of the right atrioventricular groove (Fig. 2). Further mappings were given up because of the tight operative schedule. An incision was made along the

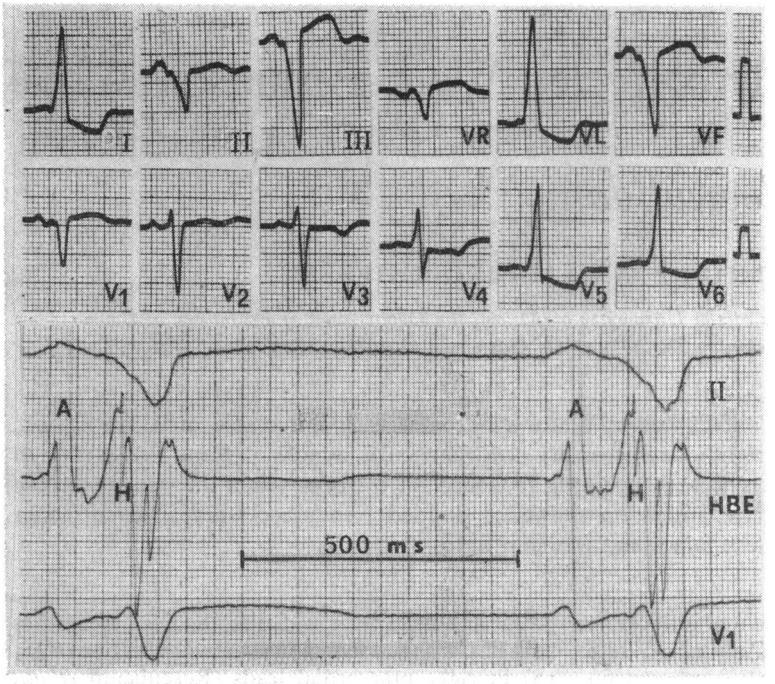

Fig. 1 (Above) *Electrocardiogram of type B WPW syndrome.* (Below) *His bundle recording, with evidence of early ventricular activation.*

lateral side of the right atrioventricular sulcus, and silk stitches were applied, together with electrocoagulation by implanted needle.

In the course of the double valvular replacement (Björk-Shiley and Smeloff-Cutter prostheses, in mitral and aortic position, respectively) the right atrium was opened, and the upper margin of the tricuspid ring incised, then sutured, leaving a 2 cm gap at the coronary sinus outlet. The electrocardiographic features of type B WPW syndrome were suppressed, but 5 days later they recurred. The heart was then re-exposed and incision was made in the right atrioventricular rim, posterolaterally, until pre-excitation and the delta wave disappeared from the electrocardiogram. Henceforth, type B WPW syndrome and related arrhythmias never recurred, the electrocardiograms only showing an impairment in the atrioventricular and right bundle-branch conduction, with inverted T waves in lead III, aVF, and from V1 to V4 (Fig. 2). The patient died from bacterial endocarditis and cardiac failure 43 days after the complete surgical correction of the syndrome.

At *necropsy*, cardiac hypertrophy and extensive pericardial adhesion from subacute pericarditis were seen. The operative fields and the valvular prostheses were regular; fine, velvety endocarditic vegetations were seen along the suture of the mitral annulus.

The *histological examination* of the conducting system of the heart and of both atrioventricular rims, on serial sections, was carried out along with the technique usually adopted by Rossi (1975). The sinoatrial node was fibrotic and marginally involved in subacute pericarditis. Gaps were present in the medial-posterior right atrioventricular ring. The atrioventricular node was normal. Atriofascicular fibres of James joined the uppermost portion of the common (His) bundle (Fig. 3). Lower Mahaim fibres anastomosed the fibrotic root of the right bundle-branch with the adjacent septal myocardium (Fig. 3). The roots of the left bundle-branch, precociously parted from the common bundle by interposition of a fibrous spur, were slightly atrophic, too. Nothing significant was noticed in the left atrioventricular rim. In the right atrioventricular groove, the subepicardial fat tissue was largely involved in postoperative scarring and fibrosing pericarditis, with patchy foreign-body (silk stitches) granulomatosis (Fig. 3), arteriolitis obliterans, and small areas of myocardial necrosis. Latero-posteriorly, close to the tricuspid insertion, on the subepicardial side, a few bundles of atrial and ventricular muscle approached one another, but were constricted and mutilated in the scarring sutures and inflammatory and fibrotic changes nearby, within the field of surgical injury (Fig. 3). The vestiges of surgically disrupted atrioventricular bundles of

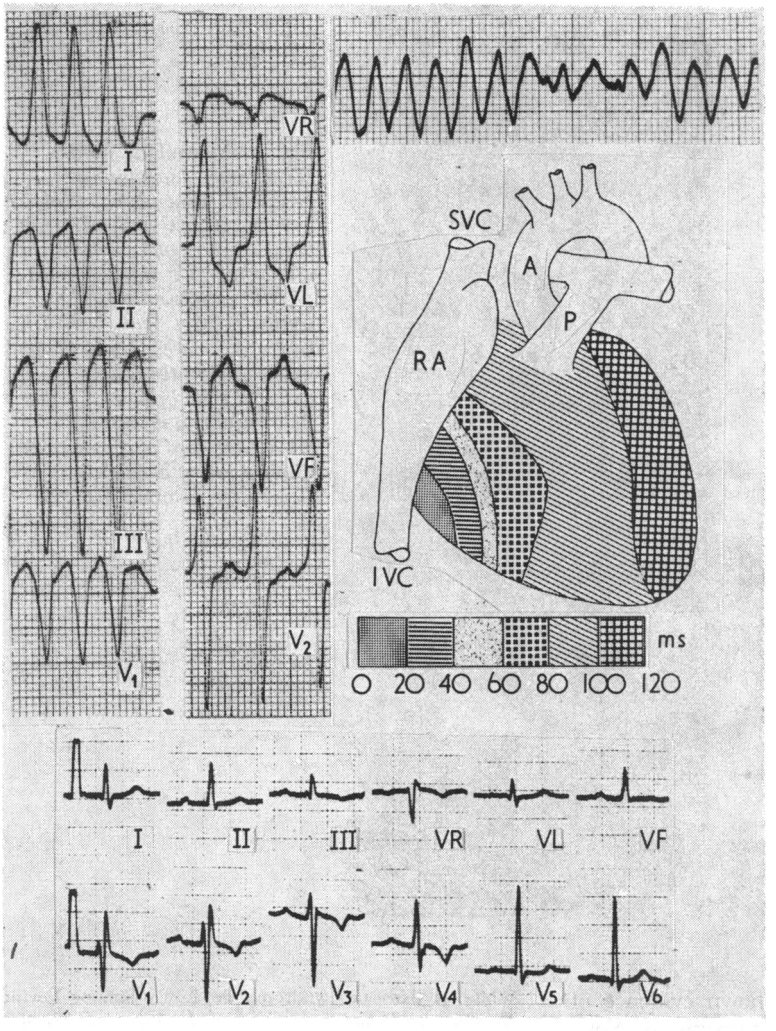

Fig. 2 Left: *atrial fibrillation with fast-rate QRS complexes showing evident delta waves.* Above right: *ventricular fibrillation.* Inset: *anterior epicardial mapping with early activation of the lateral side of the right ventricle, close to the atrioventricular groove.* Below: *after operation the electrocardiogram shows the disappearance of type B WPW syndrome.*

Kent (one or a few, close together) could hardly be expected to present themselves otherwise.

Discussion

The present data confirm the value of co-ordinated electrophysiological and surgical procedures in the diagnosis and treatment of type B WPW syndrome, particularly to prevent the high risk of intractable atrial fibrillation. Curry and Krikler (1975) and Dreifus *et al.* (1976) have stated that, when the incessant atrial impulses are conducted down to the ventricles at a very fast rate through a Kent bypass, they are liable to trigger ventricular fibrillation, as in the present case, with immediate threat to life.

Correlating the clinical and anatomical features, the complete and prolonged surgically-induced block of the anomalous atrioventricular pathway, with abolition of the WPW syndrome, can be ascribed to the division of the right-sided Kent bundle. For this very reason, however, a continuous muscle bridge across the atrioventricular sulcus could not, of course, be shown by direct histological observation, but its existence could be deduced from comparison of the present findings with the scanty microscopical evidence of Kent bundles in unoperated cases of WPW syndrome (Lev *et al.*, 1955; Verduyn Lunel, 1972; Brechenmacher *et al.*, 1974; James and Puech, 1974; Rossi, 1975; Rossi *et al.*, 1975; Dreifus *et al.*, 1976).

In the present heart atriofascicular (James) fibres and fasciculoventricular (Mahaim) fibres were

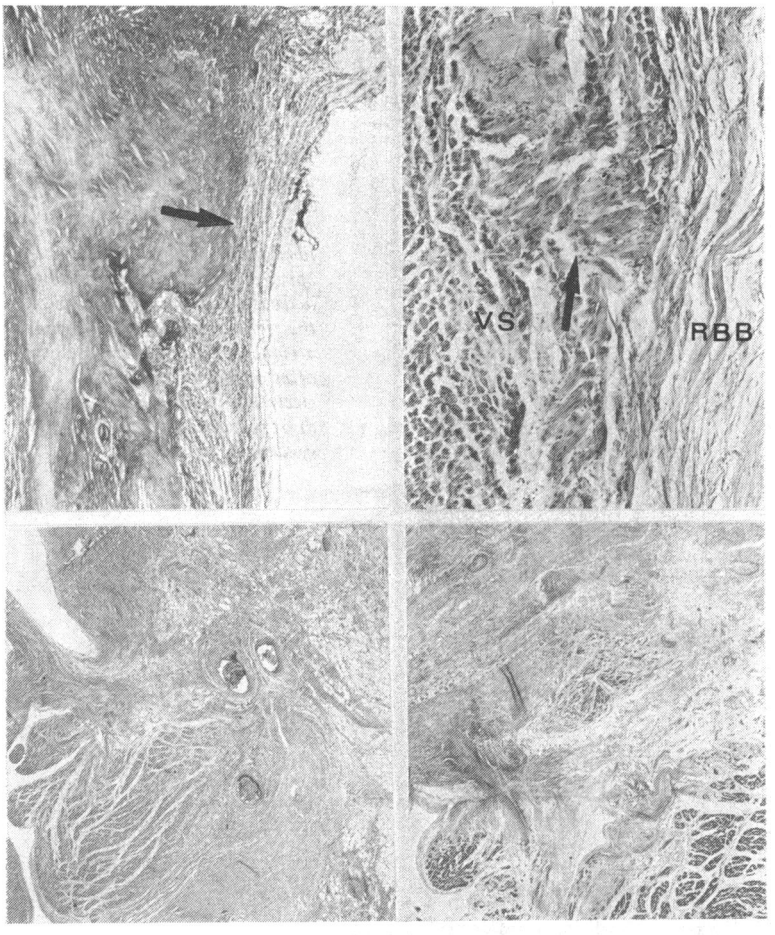

Fig. 3 Above left: *James fibres (arrow) joining the proximal common bundle. (Azan. ×7·25.)* Above right: *Mahaim fibres (arrow) anastomosing the root of the right bundle-branch (RBB) with the ventricular septum (VS) (Azan. ×72·5.)* Below left: *surgical scarring and suture close to the lateral tricuspid ring. (Azan. ×7·25.)* Below right: *a myocardial fascicle involved in the surgical scarring (Kent bundle?), close to the tricuspid ring.*

also found, but they had not shown evidence of conduction, either before or after operation. None the less, the coexistence of different anomalous atrioventricular pathways in individual cases of WPW syndrome (Rossi *et al.*, 1975; Spurrell *et al.*, 1975) has been confirmed.

As the surgical damage to the Kent bundle had been complete, and the undamaged James and Mahaim fibres apparently devoid of any functional role, one can suggest that, had the patient survived, the WPW syndrome and complicating arrhythmias would probably not have recurred.

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